

# Syringomyelia

## A patient guide

**In brief:** Syringomyelia is a long-term, slowly developing condition in which a fluid-filled cavity, called a syrinx, forms within the substance of the spinal cord and may gradually enlarge. It is almost always a secondary phenomenon — a downstream consequence of a disturbance in the flow of cerebrospinal fluid, most often at the base of the skull or within the spinal canal. Its hallmark is a distinctive, cape-like loss of pain and temperature sensation, and modern evidence places identifying and treating the underlying cause at the centre of management.

## What syringomyelia is

Syringomyelia is a disorder in which a fluid cavity develops inside the spinal cord itself. The cavity is known as a syrinx, from the Greek word for a tube or pipe. As the cavity slowly expands, it stretches and presses on the delicate nerve fibres that run through the cord, and over time this can interfere with their function. The condition is characteristically chronic and progressive rather than sudden, often evolving over years.

The spinal cord carries the pathways that transmit movement, sensation, and the control of many bodily functions between the brain and the rest of the body. A syrinx most commonly forms in the cervical region — the part of the cord within the neck — although it can extend over many segments and, in some cases, reach into the brainstem, where it is termed syringobulbia. The cavity contains fluid that is similar to, but not necessarily identical with, the cerebrospinal fluid that bathes the brain and spinal cord.

The central principle to understand about syringomyelia is that the syrinx is the lesion seen on a scan, not a complete diagnosis in itself. A cavity within the cord is a sign that something is disturbing the normal mechanics of fluid movement around and within the cord. For this reason, syringomyelia is best thought of as a signpost that points towards an underlying disturbance, and the literature of the past five decades has steadily reinforced the view that the cavity is a consequence rather than a primary disease.

## How it differs from hydromyelia

A frequent source of confusion is the distinction between syringomyelia and a closely related finding called hydromyelia. The two are not the same, and the difference carries genuine significance.

Hydromyelia describes a dilation, or widening, of the central canal of the spinal cord. The central canal is a small natural channel that runs down the middle of the cord and is in continuity with the fluid spaces of the brain, including the fourth ventricle. It is lined by a specialised layer of cells called ependyma. Hydromyelia is, in essence, an enlargement of a normal embryological space, and a small, central, ependyma-lined widening of the canal — sometimes reported as a "prominent central canal" — is frequently an incidental observation of little consequence when it is found in a person without symptoms.

A true syrinx is different. It is a cavity within the parenchyma — the working tissue — of the spinal cord. It is frequently eccentric, meaning it sits off to one side of the central canal rather than along its axis. Crucially, a true syrinx is often not lined by ependyma. Instead, it dissects through and destroys cord tissue, separating the nerve fibres and leaving a wall of reactive scar tissue, or gliosis. In other words, hydromyelia is a dilated normal structure, whereas syringomyelia is a pathological cavity that disrupts the cord itself.

Because magnetic resonance imaging often cannot fully separate the two, and because they can coexist, many authors adopt the combined term "syringohydromyelia." Classic autopsy-based research clarified that some cavities communicate with the central canal and the fourth ventricle while others do not — a distinction that has direct bearing on how a cavity is understood and approached. The practical upshot is that a small, central,

asymptomatic canal dilatation tends to be regarded as benign, whereas a true parenchymal cavity that is enlarging or producing symptoms prompts a careful search for a treatable cause.

## What causes it (Chiari, tethering, trauma, tumour) and the main theories

Syringomyelia is usually the result of another underlying condition rather than a disease that arises entirely on its own. Identifying the driver is the central task, because the cavity is the lesion and not the diagnosis. The recognised causes include the following.

**\*\*Chiari I malformation.\*\*** This is the commonest cause. In a Chiari I malformation, the lower part of the cerebellum — the cerebellar tonsils — sits lower than usual, descending five millimetres or more below the foramen magnum, the large opening at the base of the skull. This descent obstructs the free flow of cerebrospinal fluid across the foramen magnum and disturbs the pulsatile mechanics of the fluid.

**\*\*Tethered cord and spinal dysraphism.\*\*** Here the spinal cord is abnormally anchored at its lower end, altering the way the cord moves and the way fluid circulates around it.

**\*\*Post-traumatic causes.\*\*** Following a spinal cord injury, scarring of the lining around the cord (arachnoid scarring) combined with persisting narrowing of the spinal canal can obstruct the flow of fluid in the subarachnoid space. A cavity may then form at the level of the original injury and extend upwards or downwards over time. This is termed post-traumatic syringomyelia and can develop in a substantial minority of people after cord injury.

**\*\*Inflammatory arachnoiditis.\*\*** Inflammation and scarring of the arachnoid lining — which may follow infection, bleeding, surgery, or an arachnoid web, or may have no identified cause — can similarly impede fluid flow.

**\*\*Intramedullary tumour.\*\*** A tumour within the cord, such as an ependymoma or a haemangioblastoma, can be associated with a fluid cyst. Because of this, a tumour must be excluded with contrast-enhanced imaging before a cavity is labelled as having no identifiable cause.

**\*\*Idiopathic syringomyelia.\*\*** When thorough investigation, including a contrast scan of the whole neuraxis, reveals no underlying cause, the condition is described as idiopathic. This is a diagnosis of exclusion.

Several theories have been proposed over the years to explain how a cavity forms, and together they shape modern understanding. The earliest, the hydrodynamic or "water-hammer" theory, proposed that obstruction at the outlet of the fourth ventricle drives a pulse of fluid down a patent central canal. A later craniospinal pressure-dissociation theory suggested that a tonsillar "plug" acts like a valve, so that coughing or straining creates pressure differences between the head and the spine that move fluid into the cord. A pivotal advance came with the piston theory, derived from moving (cine) MRI, which showed that the tonsils move like a piston with each heartbeat, generating a pressure wave that drives fluid into the cord along the spaces around its blood vessels, without any need for a patent central canal. A further refinement, the intramedullary pulse-pressure or "Venturi" theory, proposed that the distending force is a relative rise in pressure within the cord compared with the surrounding fluid space, with a narrowing producing a suction effect that distends the cord.

The modern consensus draws these strands together: a syrinx arises from a block to fluid flow — at the foramen magnum or within the spinal subarachnoid space — that disturbs the cord's pulsatile mechanics and leads to the accumulation of fluid within the cord tissue. This is precisely why approaches that restore normal fluid flow are biologically rational.

## The characteristic symptom pattern

Syringomyelia produces a clinical signature that is, in its classic form, quite distinctive and helps to set it apart from many other spinal conditions.

Because an early syrinx expands around the central part of the cord, it first interrupts the fibres carrying pain and temperature sensation at the point where they cross from one side of the cord to the other. The fibres carrying light

touch, vibration, and the sense of position, which travel in a different part of the cord, are initially spared. The result is what is termed dissociated sensory loss: a loss of the ability to feel pain and temperature, while the sense of light touch is preserved at first.

This altered sensation classically appears across the shoulders, the upper arms, and the upper trunk, in what is often described as a "cape-like" or "shawl" distribution. Because pain and temperature are no longer felt normally in these areas, painless burns and cuts can be an early feature, as a person may not register heat or injury. Over time, other features may emerge. Wasting and weakness of the small muscles of the hands can develop, reflecting involvement of the lower motor neurons within the cord. Joints that are repeatedly injured without the protective warning of pain may become damaged and disorganised — so-called neuropathic, or Charcot, joints, classically affecting the shoulder or elbow. As the cavity enlarges, the longer nerve pathways may become involved, producing stiffness, changes in balance, and weakness in the legs. Neck and arm pain are also common. In children, a spinal curvature, or scoliosis, may be the feature that leads to the cavity being discovered.

This combination — dissociated, cape-like sensory loss, painless injuries, wasting of the hands, and the later appearance of long-tract signs — is the hallmark that distinguishes syringomyelia from most conditions that compress the cord or its nerve roots from outside.

## How it tends to present and progress

Syringomyelia typically declares itself gradually. Symptoms often begin subtly and evolve over months or years, which can make the early stages difficult to recognise. The pattern and pace of progression vary considerably between individuals and depend heavily on the underlying cause.

In Chiari-associated syringomyelia, symptoms may include headache made worse by coughing or straining, alongside the characteristic sensory and motor features as the cavity develops. In post-traumatic cases, new or changing symptoms may appear months or even years after the original spinal cord injury, sometimes signalled by a change in the level of sensory loss, new pain, or increasing weakness above the original level of injury. In children with scoliosis, the curvature itself may be the presenting feature, with the cavity found only when imaging is carried out.

It is important to note that the relationship between the size of a cavity on a scan and the severity or progression of symptoms is not straightforward. Radiological size change correlates poorly with clinical change: a cavity may remain stable in size while symptoms evolve, or may shrink without an exactly matching shift in symptoms. This is one reason why assessment combines the clinical picture with imaging rather than relying on either alone.

## How it is assessed (MRI)

Magnetic resonance imaging is the investigation of choice for syringomyelia. The standard assessment involves sagittal and axial images of the whole neuraxis — the brain and the entire length of the spine — using the sequences (T1 and T2 weighting) that show the cavity and the surrounding structures clearly.

MRI demonstrates the cavity directly, defining its size, its position within the cord, and how far it extends. The same study allows the foramen magnum to be examined, including the position of the cerebellar tonsils, so that a Chiari malformation can be identified. It also shows abnormalities of the arachnoid and dural linings that may point to scarring or a web.

The use of a contrast agent, gadolinium, is important because it helps to exclude an intramedullary tumour. A tumour-associated cyst can closely resemble other cavities on plain imaging, and contrast enhancement is the means by which a tumour is sought before a cavity is labelled as idiopathic.

A specialised technique known as cardiac-gated phase-contrast cine-MRI films the cerebrospinal fluid as it moves. By showing whether fluid is flowing freely or is obstructed at the foramen magnum and within the spinal subarachnoid space, this flow study contributes to understanding the mechanism in an individual case and to

surgical planning and selection. Taken together, these elements of an MRI examination allow the cavity, its likely cause, and the state of fluid flow to be assessed on a single study.

## How it is distinguished from related conditions

Distinguishing syringomyelia from related conditions rests on both the clinical pattern and the imaging findings.

The first distinction is from hydromyelia, described above: a dilated, central, ependyma-lined canal differs from a true parenchymal cavity, and the two are separated as far as imaging allows, with attention to whether a cavity communicates with the central canal and fourth ventricle.

The second is the separation of the syrinx from the condition that is causing it. A cavity is not the end of the diagnostic process. The presence of tonsillar descent points to a Chiari malformation; an abnormally low, anchored cord suggests tethering; a history of injury with localised scarring suggests a post-traumatic cause; inflammatory scarring of the arachnoid suggests arachnoiditis; and contrast enhancement points to a tumour. Only when a contrast study of the whole neuraxis is normal is the term idiopathic applied.

The third distinction is from conditions that compress the cord or nerve roots from outside, such as degenerative narrowing of the spine. The dissociated, cape-like sensory loss of syringomyelia, with preserved light touch early on, together with painless injuries and wasting of the hand muscles, forms a pattern that differs from the sensory and motor changes typically produced by external compression. Classification frameworks developed over the years — one based on the underlying pathology and another organised around the fluid-pathway problem to be addressed — both serve to place a given cavity within this wider context.

## What the evidence shows about management (treating the cause; shunting as last resort)

Because syringomyelia is usually driven by an underlying problem, the published evidence consistently points to one organising principle: the most durable approach is to address the underlying cause rather than the cavity itself. When the disturbance of fluid flow is corrected, the cavity frequently becomes smaller on follow-up imaging.

The way this principle is applied depends on the cause. Where a Chiari I malformation is responsible, the operation used is a decompression at the foramen magnum to create space and restore fluid flow. The literature has examined in detail whether this is best performed with or without an enlargement of the dural lining (a duraplasty). Decompression with duraplasty has been associated with better resolution of the cavity and symptoms and lower rates of repeat surgery when a syrinx is present, although with a greater chance of fluid-related complications; bony decompression alone carries fewer complications but a higher chance of recurrence. Meta-analyses have tended to favour including a duraplasty when a syrinx is present.

Where a tethered cord is responsible, the corresponding approach is to release the tethering. In post-traumatic and arachnoiditis-associated cases, the principle is to decompress and restore fluid flow — through measures such as freeing the scarred arachnoid and enlarging the dural space — rather than draining the cavity first. The outlook in these cases depends substantially on the extent of scarring: focal, localised scarring does considerably better than extensive scarring. Where a tumour is responsible, treating the tumour typically allows the associated cyst to resolve.

For idiopathic cavities found by chance and not causing symptoms, the published experience is reassuring. Most such cavities remain stable or shrink over time, and most people remain stable or improve. The evidence-supported default in this situation is careful monitoring with periodic MRI rather than intervention, with closer attention reserved for progressive symptoms.

Operating directly to drain the cavity — placing a shunt, such as a syngo-subarachnoid, syngo-peritoneal, or syngo-pleural shunt — is generally regarded as a last resort. Critical appraisals and modern systematic reviews show high long-term rates of failure and of further surgery, with cavities re-expanding in a substantial proportion

of older series and recurrent tethering being a recognised problem. For these reasons, shunting tends to be reserved for situations where the underlying cause cannot be corrected, or where cause-directed surgery has not succeeded, rather than being used as a first measure.

## Outlook

The outlook in syringomyelia varies according to the cause, the extent of any scarring, and how far the condition has progressed. Several broad themes emerge from the literature. Idiopathic cavities that are found incidentally and cause no symptoms tend to follow a favourable course, frequently remaining stable or shrinking, with reassuring long-term follow-up reported in children in particular. Where a correctable cause such as a Chiari malformation or a tethered cord is identified and addressed, the cavity often diminishes and symptoms may stabilise or improve. In post-traumatic and arachnoiditis-associated cases, focal scarring carries a better prospect than extensive scarring. A recurring observation across the evidence is that change in the size of a cavity on imaging does not map neatly onto change in symptoms, so outlook is judged on the overall clinical and radiological picture rather than on scan appearances alone. In children, identifying and treating an underlying cause may, in some circumstances, halt or even reverse an associated spinal curvature, which is one reason imaging of the neuraxis is given attention in the assessment of certain curves.